Chapter 12
Clinical Staging

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The problem of the staging of lymphedema is a perennial topic for discussion at consensus meetings within national and international congresses. First of all, for definitions and scope of pathology to be universally accepted, the requirements of simplicity, recognizability, and worldwide utilization must be met.

Four proposals based on different clinical and instrumental aspects of pathology have been presented at the international level, yet only some of the attributes are common to all. Through the work of a special world commission, a synthesis of the different proposals will provide scientific communication with universally recognized and accepted parameters.

Primary and secondary lymphedemas have different clinical stages of evolution, in part mutually reversible, that influence affected patients differently from the physical, emotional, and psychological points of view.

Achieving common acceptance of stages of lymphedema, as in other diseases, seems to be a problem that cannot be postponed further for reasons of "scientific communication" and for the undoubted medicolegal and social impact. In more advanced clinical stages, the condition takes on the characteristics of a real "social disease," the costs of which are generated both from medical care and from loss of productive capacity.

The clinical staging, reported in the "consensus document" of the International Society of Lymphology,[1] currently includes four clinical stages (Table 12.1); it initially included three clinical stages (I, II, and III), but recently, motivated by our Italian classification,[2-5] which underscored the importance of including the "preclinical" aspect of the primary and secondary types of lymphedema, potentially progressive (e.g., mastectomy with coincident limbs), the pre-clinical stage, defined

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Table 12.1 Staging according to the "consensus document" of the International Society of Lymphology

Clinical stage	Evidence
0	Subclinical with possible clinical evolution
I	Edema regressing with treatments with positive pitting test
II	Edema partially regressing with treatments with negative pitting test
III	Elephantiasis with cutaneous complications and recurrent infections

as stage 0, was included. Stage 0 refers to a latent or sub-clinical condition, where swelling is not evident despite impaired lymph transport. Stage I represents an early accumulation of fluid relatively high in protein content (in contrast to "venous edema") that subsides with limb elevation. Pitting may occur. Stage II signifies that limb elevation alone rarely reduces tissue swelling and that pitting is manifest. Stage III encompasses lymphostatic elephantiasis where pitting is absent and trophic skin changes, such as acanthosis, fat deposits, and warty overgrowths develop. The severity of the stages is based on the volume differences: minimal (<20% increase), moderate (20–40% increase), and severe (>40% increase). These stages only refer to the physical condition of the extremities.

Some healthcare workers examining disability utilize the World Health Organization's guidelines for the International Classification of Functioning, Disability, and Health (ICF). Quality of Life issues (social, emotional, physical inabilities, etc.) may also be addressed by individual clinicians and can have a favorable impact on therapy and compliance.[6]

At the recent XX World Congress of the International Society of Lymphology, held in Brazil, in a special Consensus session, a special world commission was organized to finalize a new, official staging of the International Society.[2]

In Germany, led by Prof. Ethel Foeldi, four clinical stages[11] have been introduced for the first time, adding to those reported in the actual 'consensus document' a stage 0, which represents all cases of sub-clinical lymphedema, but with a significant risk of clinical progression (e.g., lymphoscintigraphy strongly predictive; Table 12.2).

Since 1994,[2] five clinical stages have been recognized in Italy (Table 12.3). This system emphasizes the importance of pre-clinical cases at risk of evolution (in stage I) and cases of elephantiasis with major chronic inflammatory and infectious complications and risk of neoplastic tissue degeneration (stage V). Depending on the stage it is also possible to direct the therapeutic treatment toward the corresponding preventive options.[7]

In Japan, a team led by Prof. Moriji Ohkuma, a dermatologist with heightened sensitivity to infectious complications in cutaneous and subcutaneous tissues, proposed four-phase staging involving inspection and palpation of the affected areas and of assessing the frequency of the infectious episodes and inflammatory complications; based upon the developmental stage, it is possible to obtain prognostic information (Table 12.4). This is obviously a staging with a more strictly dermatological point of view; while clinical, it is conceptually valid, since it considers some clinical and inflammatory aspects, complications that are frequently found in patients with both primary and secondary forms of lymphedema.[8,9]

Table 12.2 Staging according to the German Society of Lymphology (Prof. E. Foeldi)

Clinical stage	Evidence
0	No edema, but evidence of a risk condition for evolution
I	Edema regressing with treatments with positive pitting test
II	Edema partially regressing with treatments with negative pitting test
III	Elephantiasis with cutaneous complications and recurrent infections

Table 12.3 Staging according to the Italian Society of Lymph-Angiology (Michelini–Campisi)

Clinical stage	Evidence
I	No edema in individuals at risk (pre-clinical)
II	Edema that regresses spontaneously with elevation and with night rest
III	Edema that does not regress spontaneously, only with treatments and partially
IV	Elephantiasis (abolition of tendon and bone projections)
V	Elephantiasis complicated by cutaneous and recurrent infections and impairment of deep body structures (muscles, joints)

Table 12.4 Staging according to the Japanese Society of Lymphology (Prof. M. Ohkuma)

Clinical stage	Inspection	Palpation	Acute Dermo-epidermitis	Prognosis
I	Normal	Pitting ++	Absent	Temporary
II	Thin skin	Increase in thickness, pitting +	Absent	Permanent
III	Cutaneous lichenification	Increase in thickness, pitting –	Present	Worsening
IV	Verrucosis	Pitting absent	Very often	Worsening

The staging proposed in South America, and, in particular in Brazil by Prof. Mauro Andrade, is of substantial interest because, in addition to taking into account the importance of pre-clinical cases at risk of development of infectious and degenerative complications, it also analyzes the functional effects of edema on the limb with impairment of one, two, or three major joints (Figs. 12.1–12.5). This aspect also permits better definition of the commitments of global functional rehabilitation, the degree of care needed by the patient and the impairment in "Daily Living Activities." (Table 12.5). This classification thus utilizes both clinical and functional criteria for patient assessment.[9]

It should be emphasized, however, that the new Brazilian proposal addresses deficiencies that have been recognized in other stages so far presented.

The respective positions of the "experts" at such a delicate and transitional moment for both public and private health systems in different countries also stems from the need to redefine welfare parameters for these highly prevalent diseases.[10] At more advanced stages of disease, in fact, we can identify the extremes of a true social disease for which the health system must provide incentives and normative facilitations comparable to the other diseases for which such benefits and advantages are provided.

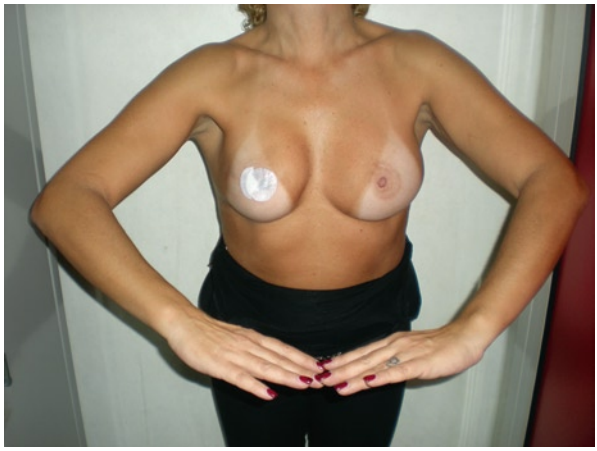

Fig. 12.1 Lymphedema stage 0 (pre-clinical)

Fig. 12.2 Lymphedema stage I (involvement of one major joint)

Fig. 12.3 Stage II (involvement of two major joints of the limb)

It is pointless to say that, currently, many National Healthcare Systems provide therapies to patients with both primary and secondary lymphedema in an inequitable manner, with poor distribution of healthcare resources. In most countries, the costs of materials, elastic garments and phlebo-lymph-active drugs are charged to the patient.

Thus, it is essential to solve these problems in each country at a governmental level. Epidemiological studies are still insufficient and must be updated in order to better define a problem that for too long has been totally ignored, while the number of patients affected is increasing daily.

It should also be noted that the various staging criteria examined take into account only aspects of the organic and physical involvement of the patients; yet, the variable emotional and psychological involvement, in some cases, regardless of the clinical evolution, the age, or the socio-economic and cultural condition of the patient, assumes greater functional significance. These factors that, over time, have a more profound influence on behavior, personal performance, and social relationships, and reinforce the simple physical problem, are overlooked in the staging schemes.

Fig. 12.4 Stage III (involvement of three major joints of the limb)

Fig. 12.5 Stage IV (cutaneous infections and inflamatory complications of the limb)

Table 12.5 Staging according to the Brazilian Society of Lymphology (Prof. M. Andrade)

Clinical stage	Evidence
0 (Fig. 12.1)	No edema in individuals at risk (pre-clinical)
I (Fig. 12.2)	Edema that regresses spontaneously with elevation, pitting ++, Stemmer +, involvement of one major joint of the limb
II (Fig. 12.3)	Edema that does not regress spontaneously, only with treatments, pitting +, Stemmer ++, involvement of at least two major joints of the limb
III (Fig. 12.4)	Edema that does not regress spontaneously, only with treatments, pitting +, Stemmer ++, involvement of three major joints of the limb
IV (Fig. 12.5)	Edema that does not regress spontaneously, only with treatments, pitting +, Stemmer ++, involvement of three major joints of the limb, with cutaneous infections

References

1. International Society of Lymphology. The diagnosis and treatment of peripheral lymphedema. Consensus document of the International Society of Lymphology. *Lymphology*. 2003;36: 84-91.
2. Michelini S, Failla A. Linfedemi: inquadramento diagnostico clinico e strumentale. *Minerva Cardioangiol*. 1997;45(6 suppl I):11-15.
3. Michelini S, Campisi C, Ricci M, et al. Linee Guida Italiane sul Linfedema. *Eur Med Phys*. 2007;43(suppl 1–3):34-41.
4. Campisi C, Michelini S, Boccardo F. Guidelines of the Società Italiana di Linfangiologia. *Lymphology*. 2004;37(4):174-179.
5. Campisi C, Michelini S, Boccardo F. Lymphology in medical and surgical practice. Italian guidelines looking towards Europe and World. *Eur J Lymphol*. 2004;14(42):26-28.
6. World Health Organization. *International Classification of Functionng, Disability and Health*. Geneva: WHO; 2001.
7. Campisi C, Michelini S, Boccardo F, Zilli A, Borrelli V. Modern stadiation of lymphedema and corresponding preventive options. *Eur J Lymphol*. 1999;7(25):27-31.
8. Ohkuma M. In lymphedema-related acute dermatitis. Abstract book of XX Congress of International Society of Lymphology. Sociedade Brasileira de Linfologia. Salvador Bahia; September 2005: 46.
9. Michelini S, Campisi C, Failla A, et al. Staging of lymphedema: comparing different proposals. *Eur J Lymphol*. 2006;16(46):7-10.
10. Tretbar LL, Morgan CL, Lee BB, Simonian SJ, Blondeau B. *Lymphedema Diagnosis and Treatment*. London: Springer; 2008.
11. Foldi M, Foldi E, Kubik S. *Foldi's Textbook of Lymphology*. Munchen: Elsevier; 2004.